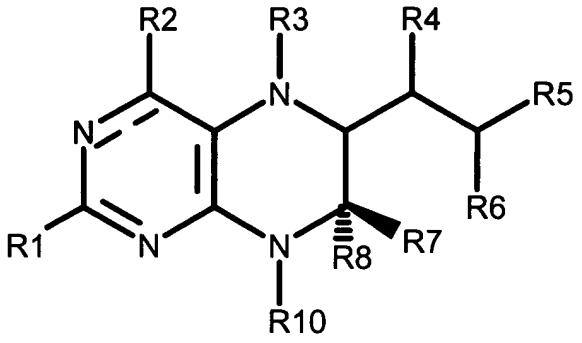
Claims

1. Use of at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

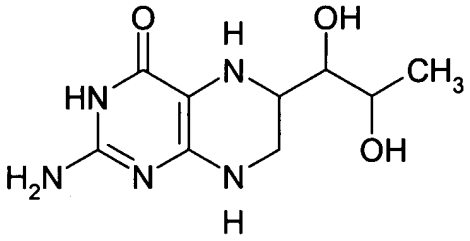
wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond; as well as their pharmaceutically acceptable salts;

for preparation of a medicament for improving protein tolerance for treatment of illnesses as a consequence of a defective amino acid metabolism.

2. Use according to Claim 1, thereby characterized, that the compound is selected from the group consisting of: 5,6,7,8- tetrahydrobiopterine, sapropterin, in particular its hydrochloride or sulfate, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

in particular there dihydrochlorides or sulfates and/or
2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

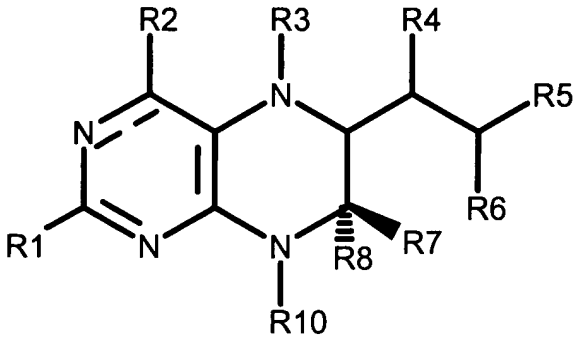
3. Use according to Claim 1 or 2, thereby characterized, that as the salts one selects hydrochloride or sulfate.

4. Use according to one of Claims 1 through 3, thereby characterized, that the amino acid metabolic disturbances include: conditions with elevated phenylaline or reduced tyrosine in body fluids, tissues or cells, in particular conditions with reduced phenylalanine hydroxylase activity, in particular conditions caused by reduced cellular access to

catecholamines, in particular orthostatic hypotension (Shy-Drager Syndrome), muscular dystonia; as well as neurotransmitter disturbances, in particular schizophrenia; phenylketonurea, in particular mild phenylketonurea, classical phenylketonurea; pigment disturbances of the skin, in particular vitiligo.

5. Use according to one of Claims 1 through 4, thereby characterized, that as the pharmaceutically acceptable salt one employs a hydrochloride.

6. Use of at least one compound with the following general formula as chaperone:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

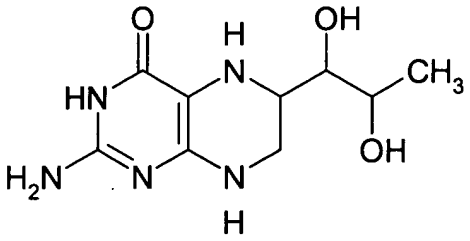
wherein R4 and R6 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond; as well as their pharmaceutically acceptable salts.

7. Use according to Claim 6, thereby characterized, that the compound is selected from the group consisting of: 5,6,7,8- tetrahydrobiopterine, sapropterin, in particular its hydrochloride or sulfate, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-Amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

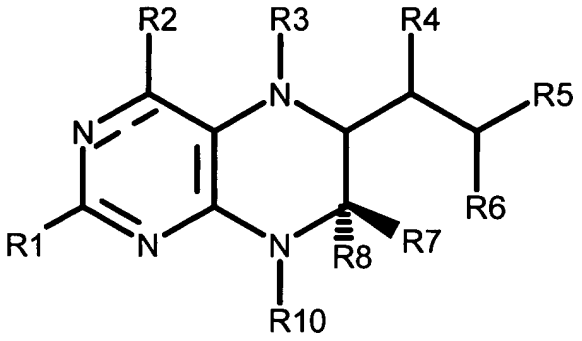
in particular their dihydrochlorides or sulfates and/or
2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

8. Use according to Claims 6 or 7 for improvement of protein mis-folding, in particular in structural anomolies in enzymes which require tetrahydrobiopterine as co-factor.

9. Use according to one of Claims 6 through 8, thereby characterized, that the enzymes were selected from: phenylalanine hydroxylase, tyrosinhydroxylase, tryptophanhydroxylase or NO-Synthase.

10. Use according to one of Claims 6 through 8, thereby characterized, that the chaperone is used as neurotransmitter and/or second messenger enhancer, in particular is suited for therapy of conditions with elevated phenylalanine or reduced tyrosin, serotonin, or dopamine in body fluids, tissues or cells, in particular in conditions with reduced phenylalanine hydroxylase, tyrosinhydroxylase, tryptophanhydroxylase or NO-Synthase activity can be employed.

11. Use of at least one compound according to the following general formula as neurotransmitter or as second messenger enhancer, in particular for catecholomine and/or serotonin or dopamine and/or nitrous oxide (NO):



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

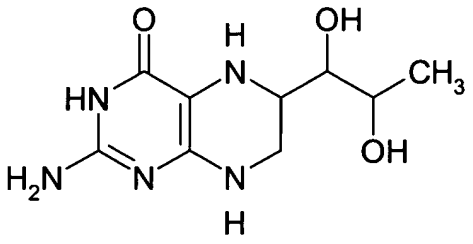
wherein R4 and R6 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond; as well as their pharmaceutically acceptable salts.

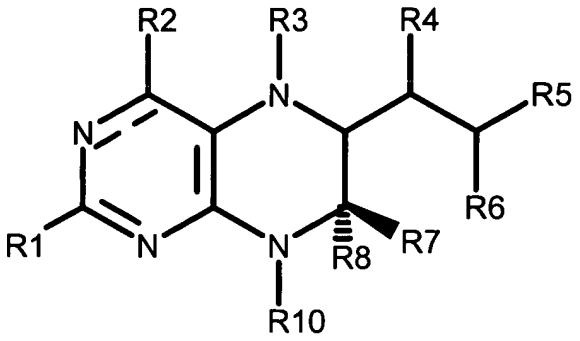
12. Use according to Claim 11, thereby characterized, that the compound is selected from the group consisting of: 5,6,7,8- tetrahydrobiopterine, sapropterine, in particular its hydrochloride or sulfate, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

in particular their dihydrochlorides or sulfates and/or
2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

13. Composition containing at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond; as well as their pharmaceutically acceptable salts; as well as

at least one amino acid, selected from the group consisting of the essential amino acids: isoleucine, leucine, lysine, methionine, threonine, tryptophane, valine, histidine; as well

as from the non-essential amino acids, in particular alanine, arginine, asparaginic acid, asparagine, cysteine, in particular acetylcysteine, glutamic acid, glutamine, clycine, proline, serine as well as tyrosine.

14. Composition according to Claim 13, thereby characterized, that it contains the essential amino acids, select from the group consisting of isoleucine, leucine, lysine, methionine, threonine, tryptophane, valine, histidine and supplementally at least one of the amino acids alanine, arginine, asparaginic acid, asparagine, cysteine, in particular acetylcysteine, glutamic acid, glutamine, clycine, proline, serine as well as tyrosine.

15. Composition according to Claim 13 or 14, thereby characterized, that it further contains carbohydrates, in particular glucose and/or vitamins.

16. Composition according to one of Claims 13 through 15, thereby characterized, that it is formulated as a preparation to be administered orally or intravenously.

17. Composition according to Claim 16, thereby characterized, that the preparation is formulated in the form of a powder, tablet, capsule, pill, in droplet form or formulated for topical application, in particular salves; as well as solutions for intravenous administration.

18. Composition according to one of Claims 13 through 17, thereby characterized, that it is in the form of a pharmaceutical composition, in certain cases with pharmaceutical galenic conventional additives.

19. Composition according to one of Claims 13 through 18, thereby characterized, that it is in the form of a dietetic composition, in certain cases with consumable additives, in particular emulsifiers, preferably lecithin, choline.

20. Composition according to one of Claims 13 through 19, thereby characterized, that it further includes minerals and/or electrolytes, which are selected from: mineral salts;

saline salts; sea salts, trace elements, in particular selenium, manganese, copper, zinc, molybdenum, iodide, chrome; alkali ions, in particular lithium, sodium, potassium; earth alkali ions, in particular magnesium, calcium; iron.

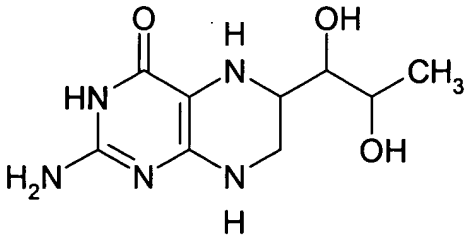
21. Composition according to one of Claims 13 through 20, thereby characterized, that it further includes phenylalanine.

22. Composition according to one of Claims 13 through 21, thereby characterized, that it further includes L-carnitine.

23. Composition according to one of Claims 13 through 22, thereby characterized, that it further includes myoinositol and choline.

24. Composition according to one of Claims 13 through 23, thereby characterized, that it contains antioxidants, in particular vitamin C.

25. Composition according to one of Claims 13 through 24, thereby characterized, that the composition is selected from the group consisting of: 5,6,7,8- tetrahydrobiopterine, sapropterin, in particular its hydrochloride or sulfate, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

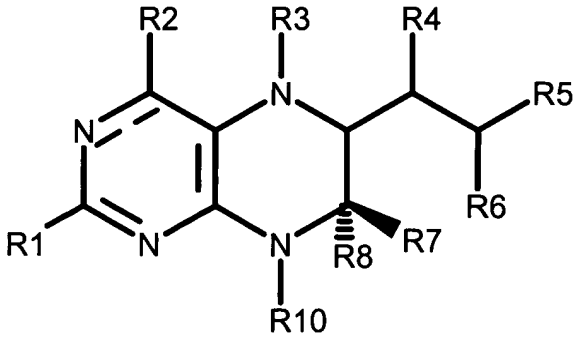
in particular their dihydrochlorides or sulfates and/or

2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

26. Use of at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

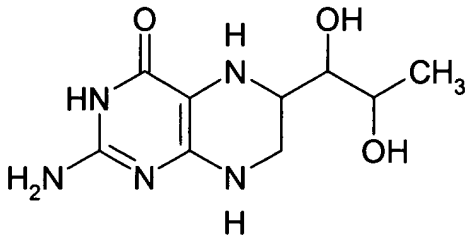
wherein R4 and R6 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

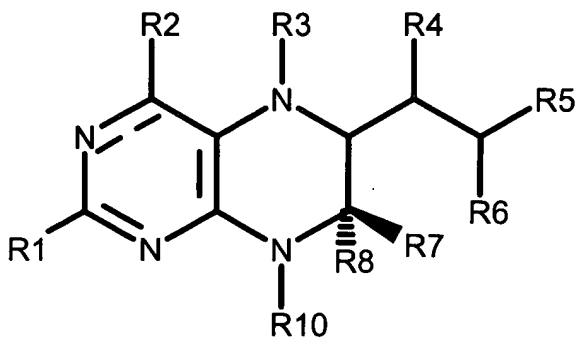
wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond; as well as their suitable salts, as nutrient supplements.

27. Use according to Claim 26, thereby characterized, that a compound is selected from a group consisting of: 5,6,7,8- tetrahydrobiopterine, sapropterin, in particular its hydrochloride or sulfate, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,
in particular their dihydrochlorides or sulfates and/or
2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

28. Special nutrient based upon essentially phenylalanine-free amino acid mixtures, thereby characterized, that it contains at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

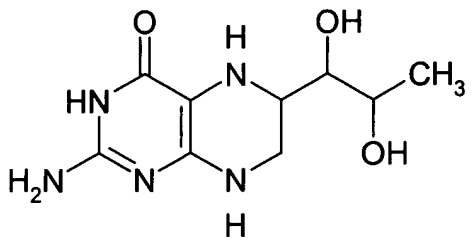
wherein R4 and R6 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond, as well as their food or comestible acceptable salts.

29. Special nutrient according to Claim 28, thereby characterized, that it contains at least one compound, which is selected from the group consisting of:
5,6,7,8- tetrahydrobiopterine, sapropterin, in particular its hydrochloride or sulfate, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,
in particular there dihydrochlorides or sulfates and/or
2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

30. Special nutrient according to Claim 28 or 29, thereby characterized, that it further contains carbohydrates, in particular glucose, maltodextrin, starches and/or fats, such as fish oil, in particular salmon oil, herring oil, mackerel oil, or tuna fish oil.

31. Special nutrient according to one of Claims 28 through 30, thereby characterized, that it is hypoallergenic and/or essentially gluten free.

32. Special nutrient according to one of Claims 28 through 31, thereby characterized, that it is an infant formula.

33. Special nutrient according to one of Claims 28 through 32, thereby characterized, that it is in the form of a powder, in particular a lyophilizate (freeze dried).

34. Special nutrient according to one of Claims 28 through 33, thereby characterized, that it further contains fatty acid supplements, in particular unsaturated fatty acids, preferably Omega-3 fatty acids, in particular alpha linolic acid, docosahexanoic acid, eicosapentanoic acid or Omege-6 fatty acids, in particular arachidonic acid, linolic acid, linolenic acid; or oleic acid.

35. Special nutrient according to one of Claims 28 through 34, thereby characterized, that it contains fish oil supplements, in particular of salmon, herring, mackerel or tuna fish oil.

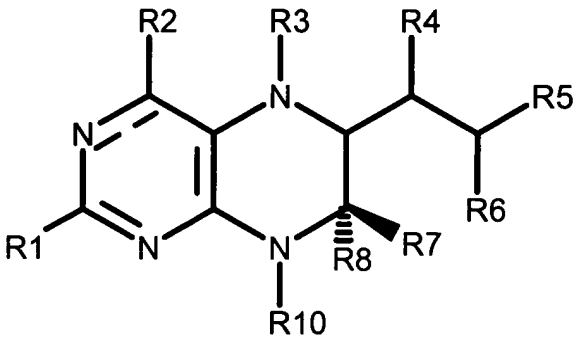
36. Special nutrient according to one of Claims 28 through 35, thereby characterized, that it is capable of use as milk substitute, in particular for infants.

37. Special nutrient or diet according to Claim 36, thereby characterized, that the milk substitute includes a fat component, wherein in particular 90% is in the form of triglycerides, 10% in the form of mono and diglycerides.

38. Special nutrient according to Claim 37, thereby characterized, that the fat component includes safflower oil and/or soy oil and/or cocoa oil.

39. Special nutrient or diet according to one of Claims 28 through 38, thereby characterized, that the milk substitute is in the form of milk mixed drink, in particular fruit milk mixed drink or cocoa.

40. Phenylalanine-poor special nutrient composition, containing a protein-poor base nutrient means as well as at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

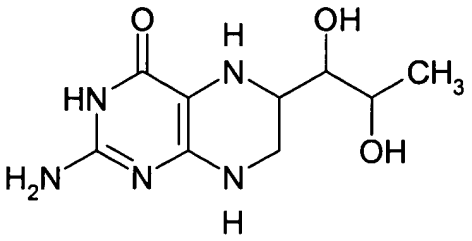
wherein R4 and R6 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond; as well as their pharmaceutically acceptable salts.

41. Phenylalanine poor special nutrient according to Claim 40, thereby characterized, that it contains at least one compound, which is select from the group consisting of: 5,6,7,8-tetrahydrobiopterine, sapropterin, in particular its hydrochloride or sulfate, as well as a compound with the following structure:

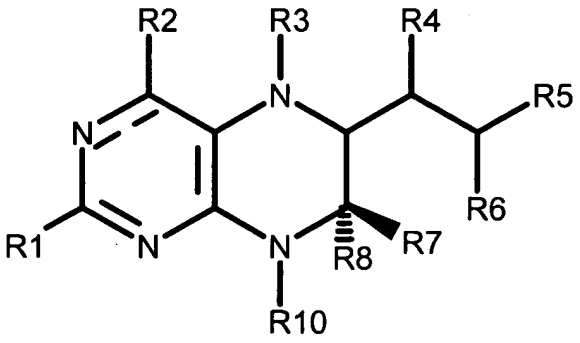


(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

in particular their dihydrochlorides or sulfates and/or
2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or
2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

42. Phenylalanine-poor special nutrient according to Claim 40 or 41, thereby characterized, that is selected from pre-cooked dishes; dough products, in particular noodles; baked products, in particular bread, cake, biscuits; sweets, in particular chocolate, candy, ice cream; drinks, in particular milk substitute means, in the form of milk mixed drink, in particular as fruit milk mix drink or cocoa; as well as beer.

43. Diagnostic test for diagnosis of tetrahydrobiopterine sensitivity in diseases of amino acid metabolism derived from, containing at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms, in particular CH_3O, preferably 9 to 32, preferably 9 to 20 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue, in particular a C9 to C32 acyl residue, preferably a C9 to C20 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independent of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

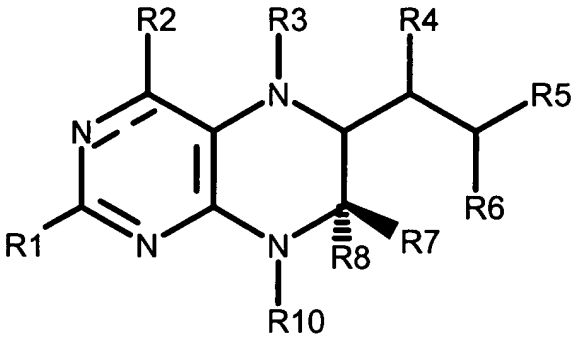
wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond; as well as their pharmaceutically acceptable salts.

44. Use according to Claim 10, thereby characterized, that the conditions include: phenylketonurea, in particular mild phenylketonurea, classical phenylketonurea; pigmentation defects of the skin, in particular vitiligo; as well as conditions caused by reduced cellular access to catecholamine, in particular orthostatic hypotension (Shy-Drager Syndrome), muscular dystonia; as well as neurotransmitter dysfunction, in particular schizophrenia; conditions caused by reduced cellular access to dopamine or serotonin as consequence of tyrosinhydroxylase, tryptophanhydroxylase deficiency in particular parkinsonionisom, depressive diseases as well as dystonia movement interference, conditions with reduced NO-Synthase activity, in particular endothelial dysfunction, immuno- deficiency.

1-44. (canceled)

45. (new) A method for long-term treatment of conditions of reduced protein tolerance due to reduced phenylalanine oxidation without deficiency of cofactor tetrahydrobiopterine, said conditions caused by mutations in the phenylalanine hydroxylase gene associated with at least one of the following allele pairs: A403V + IVS4+5G>T, P314S +R408W, F39L + D414N, Y414C + D415N, Y417H + Y417H, F55L + S310Y, V177M + R408W, P275L +Y414C, V245A +R408W, L48S + R158Q, Y417H + Y417H, V245A +R408W, R261X +A300R, R158Q + E390G, Y414C +IVS12+1G>A, I65S +A300S, H170O + A300S, R261Q + Y414C, K274fsdel11bp + E390G, IVS4-5C>G + R480W, I65T + Y414c, E390G + IVS12+1G>A, I65V + R261Q, R158Q + Y414C,

said method comprising administering a medicament containing at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

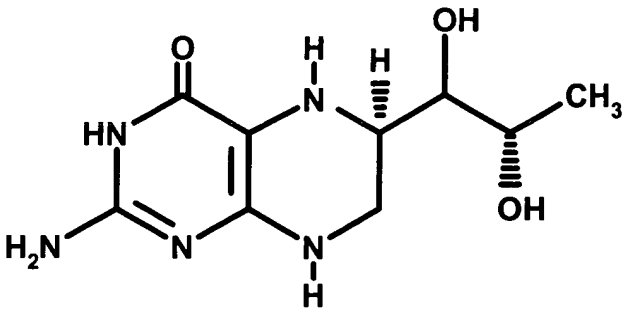
wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond;

as well as their pharmaceutically acceptable salts.

46. (new) A method as in claim 45, wherein said medicament is administered to a patient in need thereof until said patient exhibits improvement in protein tolerance.

47. (new) A method as in claim 45, wherein R1 is NH-acyl, wherein the acyl residue contains CH_3O or 9 to 32 carbon atoms, and wherein at least one of R4 and R6 are C9 to C32 acyl residue.

48. (new) A method according to Claim 45, wherein the compound is selected from the group consisting of: 5,6,7,8- tetrahydrobiopterine, sapropterin, a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

and/or

2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-etrahydrobiopterine; and/or

2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

49. (new) A method according to claim 45, wherein said pharmaceutically acceptable salt is a hydrochloride or a sulphate.

50. (new) A method according to Claim 45, wherein the condition of reduced protein tolerance is at least one of: conditions with elevated phenylaline or reduced tyrosine in body fluids, tissues or cells.

51. (new) A method as in claim 50, wherein said condition of reduced protein tolerance is classic phenylketonurea, mild phenylketonurea, or mild hyperphenylalaninemia.

52. (new) A method according to claim 45, wherein said medicament functions as chaperone for improving protein folding, in particular in the case of structural anomalies of enzymes, which require tetrahydrobiopterine as cofactor.

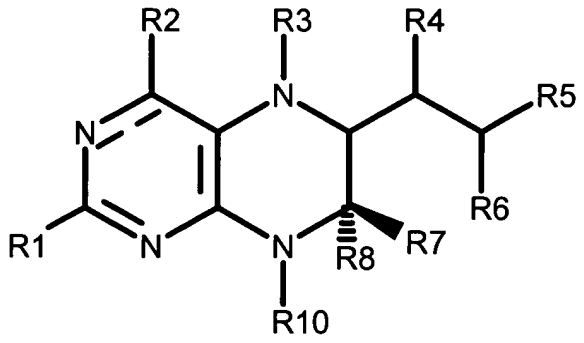
53. (new) A method according to claim 52, wherein said enzyme is selected from phenylalanine hydroxylase, tyrosinhydroxylase, tryptophanhydroxylase and NO-synthase.

54. (new) A method according to claim 45, wherein said compound functions as chaperone as neurotransmitter and/or second messenger enhancer, in particular in conditions with elevated phenylalanine or reduced tyrosin, serotonin, or dopamine in body fluids, tissues or cells, in particular in conditions with reduced phenylalanine hydroxylase, tyrosinhydroxylase, tryptophanhydroxylase or NO-synthase activity.

55. (new) A method according to claim 45, wherein said compound functions as neurotransmitter and/or second messenger enhancer, in particular for catecholamine and/or seratonin and/or dopamine and/or nitrous oxide (NO).

56. (new) A composition containing

(a) at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond;

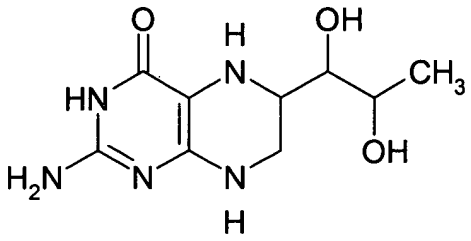
as well as their pharmaceutically acceptable salts, as well as

(b) at least one amino acid selected from the group consisting of

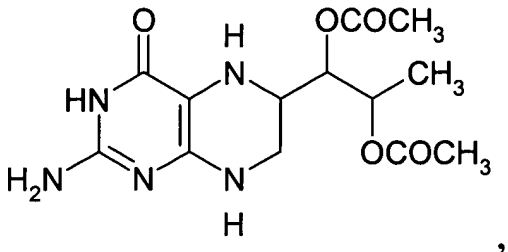
- essential amino acids: isoleucine, leucine, lysine, methionine, threonine, tryptophane, valine, histidine; as well as from

- the non-essential amino acids, in particular alanine, arginine, asparaginic acid, asparagine, cysteine, in particular acetylcysteine, glutamic acid, glutamine, clycine, proline, serine as well as tyrosine,

wherein the following compounds are excluded:



and



,

in the case that the amino acid is one of tryptophane, cysteine, in particular acetylcysteine, and tyrosine.

57. (new) A composition according to claim 56, wherein the essential amino acids are selected from the group consisting of isoleucine, leucine, lysine, methionine, threonine, tryptophane, valine, histidine; and that it further contains at least one of the following amino acids: alanine, arginine, asparaginic acid, asparagine, cysteine, in particular acetylcysteine, glutamic acid, glutamine, clycine, proline, serine as well as tyrosine.

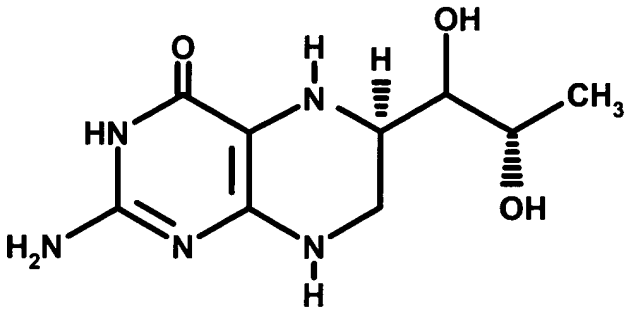
58. (new) A composition according to claim 56, further comprising a hydrocarbon, in particular glucose, and/or vitamins.

59. (new) A composition according to claim 56, formulated as an oral or intravenous preparation.

60. (new) A composition according to claim 59, wherein said composition is in the form of a powder, tablet, capsule, pill, droplets, or as solution for IV administration.

61. (new) A composition according to claim 56, in the form of a pharmaceutical preparation, optionally with pharmaceutical adjuvants or excipients.

62. (new) A composition according to claim 56, wherein the compound is selected from the group consisting of: sapropterin, in particular the hydrochloride thereof, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone, in particular the dihydrochloride or sulphate thereof,

and/or

2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-etrahydrobiopterine; and/or

2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.